Complete Summary

GUIDELINE TITLE

Posterior urethral values. In: Guidelines on paediatric urology.

BIBLIOGRAPHIC SOURCE(S)

Posterior urethral valves. In: Tekgul S, Riedmiller H, Gerharz E, Hoebeke P, Kocvara R, Nijman R, Radmayr C, Stein R. Guidelines on paediatric urology. Arnhem, The Netherlands: European Association of Urology, European Society for Paediatric Urology; 2008 Mar. p. 72-7. [30 references]

GUIDELINE STATUS

This is the current release of the guideline.

COMPLETE SUMMARY CONTENT

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SCOPE

DISEASE/CONDITION(S)

Posterior urethral valves

GUIDELINE CATEGORY

Diagnosis
Evaluation
Management
Screening
Treatment

CLINICAL SPECIALTY

Obstetrics and Gynecology
Pediatrics
Surgery
Urology

INTENDED USERS

Physicians

GUIDELINE OBJECTIVE(S)

- To outline a practical and preliminary approach to paediatric urological problems
- To increase the quality of care for children with urological problems

TARGET POPULATION

- Infants born with posterior urethral valves
- Fetuses identified antenatally with posterior urethral valves

INTERVENTIONS AND PRACTICES CONSIDERED

Diagnosis/Evaluation

1. Antenatal and postnatal ultrasonography
2. Voiding cystourethrography
3. Nuclear renography with split renal function
4. Blood tests: creatinine, blood urea nitrogen, electrolytes

Treatment/Management

1. Antenatal treatment
    - Placement of a vesicoamniotic shunt
2. Postnatal treatment
    - Bladder drainage
    - Valve ablation
    - Vesicostomy
    - High diversion
    - Life-long monitoring for bladder dysfunction, urinary infection, renal function
    - Treatment of bladder dysfunction: clean intermittent catheterization, overnight drainage, alpha-blockers, or anticholinergics for overactive bladder

MAJOR OUTCOMES CONSIDERED

- Renal function
- Bladder function
- Morbidity and mortality of treatment

METHODOLOGY

METHODS USED TO COLLECT/SELECT EVIDENCE

Searches of Electronic Databases

DESCRIPTION OF METHODS USED TO COLLECT/SELECT THE EVIDENCE

The guidelines were based on current literature following a systematic review using MEDLINE.

NUMBER OF SOURCE DOCUMENTS

Not stated

METHODS USED TO ASSESS THE QUALITY AND STRENGTH OF THE EVIDENCE

Weighting According to a Rating Scheme (Scheme Given)

RATING SCHEME FOR THE STRENGTH OF THE EVIDENCE

Levels of Evidence

1a Evidence obtained from meta-analysis of randomized trials

1b Evidence obtained from at least one randomized trial

2a Evidence obtained from at least one well-designed controlled study without randomization

2b Evidence obtained from at least one other type of well-designed quasi-experimental study

3 Evidence obtained from well-designed non-experimental studies, such as comparative studies, correlation studies and case reports

4 Evidence obtained from expert committee reports or opinions or clinical experience of respected authorities

METHODS USED TO ANALYZE THE EVIDENCE

Systematic Review

DESCRIPTION OF THE METHODS USED TO ANALYZE THE EVIDENCE

Application of a structured analysis of the literature was not possible due to a lack of well-designed studies. Whenever possible, statements have been classified in terms of level of evidence and grade of recommendation. Due to the limited

availability of large randomized controlled trials – influenced also by the fact that a considerable number of treatment options relate to surgical interventions on a large spectrum of different congenital problems – this document is therefore largely a consensus document.

METHODS USED TO FORMULATE THE RECOMMENDATIONS

Expert Consensus

DESCRIPTION OF METHODS USED TO FORMULATE THE RECOMMENDATIONS

- The first step in the European Association of Urology (EAU) guidelines procedure is to define the main topic.
- The second step is to establish a working group. The working groups comprise about 4-8 members, from several countries. Most of the working group members are academic urologists with a special interest in the topic. In general, general practitioners or patient representatives are not part of the working groups. A chairman leads each group. A collaborative working group consisting of members representing the European Society for Paediatric Urology (ESPU) and the EAU has gathered in an effort to produce the current update of the paediatric urology guidelines.
- The third step is to collect and evaluate the underlying evidence from the published literature.
- The fourth step is to structure and present the information. The strength of the recommendation is clearly marked in three grades (A-C), depending on the evidence source upon which the recommendation is based. Every possible effort is made to make the linkage between the level of evidence and grade of recommendation as transparent as possible.

RATING SCHEME FOR THE STRENGTH OF THE RECOMMENDATIONS

Grades of Recommendation

A. Based on clinical studies of good quality and consistency addressing the specific recommendations and including at least one randomized trial
B. Based on well-conducted clinical studies, but without randomized clinical studies
C. Made despite the absence of directly applicable clinical studies of good quality

COST ANALYSIS

A formal cost analysis was not performed and published cost analyses were not reviewed.

METHOD OF GUIDELINE VALIDATION

Internal Peer Review

DESCRIPTION OF METHOD OF GUIDELINE VALIDATION

There is no formal external review prior to publication.

The Appraisal of Guidelines for Research and Evaluation (AGREE) instrument was used to analyse and assess a range of specific attributes contributing to the validity of a specific clinical guideline.

The AGREE instrument, to be used by two to four appraisers, was developed by the AGREE collaboration (www.agreecollaboration.org) using referenced sources for the evaluation of specific guidelines. (See the "Availability of Companion Documents" field for further methodology information).

RECOMMENDATIONS

MAJOR RECOMMENDATIONS

Diagnosis

An obstruction above the level of the urethra affects the whole urinary tract in varying degrees.

- The prostatic urethra is distended and the ejaculatory ducts may be dilated due to urinary reflux. The bladder neck is hypertrophied and rigid.
- The hypertrophied bladder occasionally has multiple diverticula.
- Nearly all valve patients have dilatation of both upper urinary tracts. This may be due to the valve itself and the high pressure in the bladder, or due to obstruction of the ureterovesical junction by the hypertrophied bladder.
- If there is secondary reflux, the affected kidney functions poorly in most cases.

During prenatal ultrasonography screening, bilateral hydroureteronephrosis and a distended bladder are suspicious signs of a urethral valve. If a dilated posterior urethra and a thick-walled bladder ('keyhole' sign) are seen, a posterior urethral valve (PUV) is likely. In the presence of increased echogenity of the kidney, dilatation of the urinary tract and oligohydramnion, the diagnosis of a PUV should strongly be considered.

A voiding cystourethrography (VCUG) confirms a PUV diagnosis. This study is essential whenever there is a question of an infravesical obstruction, as the urethral anatomy is well outlined during voiding. A secondary reflux is observed in at least 50% of patients with PUV. Reflux is consistently associated with renal dysplasia in patients with PUV. It is generally accepted that reflux in the renal units acts as a 'pressure pop-off valve', which would protect the other kidney, leading to a better prognosis. Other types of pop-off mechanism include bladder diverticula and urinary extravasation, with or without urinary ascites. However, in the long-term, a supposed protective effect did not show a significant difference compared to other patients with PUV.

Nuclear renography with split renal function is important to assess kidney function. Creatinine, blood urea nitrogen and electrolytes should be monitored closely during the first few days. A nadir creatinine of 80 micromol/L is correlated with a better prognosis.

Treatment

Antenatal Treatment

About 40-60% of PUV are discovered before birth. The intrauterine obstruction leads to a decreased urine output, which could result in an oligohydramnios. Amnion fluid is necessary for normal development of the lung and its absence may lead to pulmonary hypoplasia, causing a life-threatening problem. Intrauterine attempts have been made to treat a fetus with PUV.

As renal dysplasia is not reversible, it is important to identify those fetuses with good renal function. A sodium level below 100 mmol/L, a chloride value of <90 mmol/L and an osmolarity below 200 mOsm/L found in three fetal urine samples gained on three different days are associated with a better prognosis.

The placing of a vesicoamniotic shunt has a complication rate of 21-59%, dislocation of the shunt occurs in up to 44%, mortality lies between 33% and 43%, and renal insufficiency is above 50%. Although shunting is effective in reversing oligohydramnios, it makes no difference to the outcome and long-term results of patients with PUV.

Postnatal Treatment

Bladder drainage. If a boy is born with suspected PUV, drainage of the bladder and, if possible, an immediate VCUG is necessary. A neonate can be catheterized with a 3.5-5 F catheter. A VCUG is performed to see if the diagnosis is correct and whether the catheter is within the bladder and not in the posterior urethra. An alternative option is to place a suprapubic catheter, perform a VCUG and leave the tube until the neonate is stable enough to perform an endoscopic incision or resection of the valve.

Valve ablation. When the medical situation of the neonate has stabilized and the creatinine level decreased, the next step is to remove the intravesical obstruction. Small paediatric cystoscopes and resectoscopes are now available either to incise or to resect the valve at the 4-5, 7-8 or 12 o'clock position, or at all three positions, depending on the surgeon's preference. It is important to avoid extensive electrocoagulation as the most common complication of this procedure is stricture formation.

Vesicostomy. If the child is too small and/or too ill to undergo endoscopic surgery, a vesicostomy is used to drain the bladder temporarily. If initially a suprapubic tube has been inserted, this can be left in place for 6-12 weeks. Otherwise, a cutaneous vesicostomy provides an improvement or stabilization of upper urinary tracts in over 90% of cases. Although there has been concern that a vesicostomy could decrease bladder compliance or capacity, so far there are no valid data to support these expectations.

High diversion. If bladder drainage is insufficient to drain the upper urinary tract, high urinary diversion should be considered. Diversion may be suitable if there are recurrent infections of the upper tract, no improvement in renal function and/or an increase in upper tract dilatation, despite adequate bladder drainage. The

choice of urinary diversion depends on the surgeon's preference for high loop ureterostomy, ring ureterostomy, end ureterostomy or pyelostomy, with each technique having advantages and disadvantages. Reconstructive surgery should be delayed until the upper urinary tract has improved as much as can be expected.

Reflux is very common in PUV patients (up to 72%) and it is described bilaterally in up to 32%. High-grade reflux is mostly associated with a poor functioning kidney. However, early removal of the renal unit seems to be unnecessary, as long as it causes no problems. It may be necessary to augment the bladder and in this case the ureter may be used.

Life-long monitoring of these patients is mandatory, as bladder dysfunction is not uncommon and the delay in day- and night-time continence is a major problem. Poor bladder sensation and compliance, detrusor instability and polyuria (especially at night) and their combination are responsible for bladder dysfunction. Between 10% and 47% of patients may develop end-stage renal failure. Renal transplantation in these patients can be performed safely and effectively. Deterioration of the graft function is mainly related to lower urinary tract dysfunction.

CLINICAL ALGORITHM(S)

The original guideline document contains an algorithm providing information on assessment, treatment and follow up of newborns with possible posterior urethral valves.

EVIDENCE SUPPORTING THE RECOMMENDATIONS

TYPE OF EVIDENCE SUPPORTING THE RECOMMENDATIONS

The type of supporting evidence is not specifically stated for each recommendation.

BENEFITS/HARMS OF IMPLEMENTING THE GUIDELINE RECOMMENDATIONS

POTENTIAL BENEFITS

- Appropriate and timely diagnosis, treatment, and management of posterior urethral valve in neonates and children
- Improvement in urinary tract and renal function

POTENTIAL HARMS

- The placing of a vesicoamniotic shunt has a complication rate of 21% to 59%, dislocation of the shunt occurs in up to 44%, mortality lies between 33% and 43%, and renal insufficiency is above 50%.
- Stricture formation is the most common complication of valve ablation procedures in which electrocoagulation is used.

QUALIFYING STATEMENTS

QUALIFYING STATEMENTS

The purpose of these texts is not to be proscriptive in the way a clinician should treat a patient but rather to provide access to the best contemporaneous consensus view on the most appropriate management currently available. European Association of Urology (EAU) guidelines are not meant to be legal documents but are produced with the ultimate aim to help urologists with their day-to-day practice.

IMPLEMENTATION OF THE GUIDELINE

DESCRIPTION OF IMPLEMENTATION STRATEGY

The European Association of Urology (EAU) Guidelines long version (containing all 19 guidelines) is reprinted annually in one book. Each text is dated. This means that if the latest edition of the book is read, one will know that this is the most updated version available. The same text is also made available on a CD (with hyperlinks to PubMed for most references) and posted on the EAU websites Uroweb and Urosource (www.uroweb.org/professional-resources/guidelines/ & http://www.urosource.com/diseases/).

Condensed pocket versions, containing mainly flow-charts and summaries, are also printed annually. All these publications are distributed free of charge to all (more than 10,000) members of the Association. Abridged versions of the guidelines are published in European Urology as original papers. Furthermore, many important websites list links to the relevant EAU guidelines sections on the association websites and all, or individual, guidelines have been translated to some 15 languages.

IMPLEMENTATION TOOLS

Clinical Algorithm

For information about availability, see the "Availability of Companion Documents" and "Patient Resources" fields below.

INSTITUTE OF MEDICINE (IOM) NATIONAL HEALTHCARE QUALITY REPORT CATEGORIES

IOM CARE NEED

Getting Better
Living with Illness

IOM DOMAIN

Effectiveness
Patient-centeredness

IDENTIFYING INFORMATION AND AVAILABILITY

BIBLIOGRAPHIC SOURCE(S)

Posterior urethral valves. In: Tekgul S, Riedmiller H, Gerharz E, Hoebeke P, Kocvara R, Nijman R, Radmayr C, Stein R. Guidelines on paediatric urology. Arnhem, The Netherlands: European Association of Urology, European Society for Paediatric Urology; 2008 Mar. p. 72-7. [30 references]

ADAPTATION

Not applicable: The guideline was not adapted from another source.

DATE RELEASED

2008 Mar

GUIDELINE DEVELOPER(S)

European Association of Urology - Medical Specialty Society
European Society for Paediatric Urology - Medical Specialty Society

SOURCE(S) OF FUNDING

European Association of Urology

GUIDELINE COMMITTEE

Not stated

COMPOSITION OF GROUP THAT AUTHORED THE GUIDELINE

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FINANCIAL DISCLOSURES/CONFLICTS OF INTEREST

All members of the working group submit a conflict of interest form. The information is kept on file in the European Association of Urology (EAU) Central Office database. This guidelines document was developed with the financial support of the EAU. No external sources of funding and support have been involved. The EAU is a non-profit organisation and funding is limited to administrative assistance, travel, and meeting expenses. No honoraria or other reimbursements have been provided.

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This is the current release of the guideline.

GUIDELINE AVAILABILITY

Electronic copies: Available in Portable Document Format (PDF) from the European Association of Urology Web site.

Print copies: Available from the European Association of Urology, PO Box 30016, NL-6803, AA ARNHEM, The Netherlands.

AVAILABILITY OF COMPANION DOCUMENTS

The following are available:

- EAU guidelines office template. Arnhem, The Netherlands: European Association of Urology (EAU); 2007. 4 p.
- The European Association of Urology (EAU) guidelines methodology: a critical evaluation. Arnhem, The Netherlands: European Association of Urology (EAU); 18 p.

Print copies: Available from the European Association of Urology, PO Box 30016, NL-6803, AA ARNHEM, The Netherlands.

PATIENT RESOURCES

None available

NGC STATUS

This NGC summary was completed by ECRI Institute on November 18, 2008. The information was verified by the guideline developer on December 19, 2008.

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